THE APPEARANCE OF NEUROBLASTOMA IN CT

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ABSTRACT

CT scans were reviewed of 22 children diagnosed with neuroblastoma but not yet treated. Most of the children had abdominal mass or bone pain. 63.6% (14/22) of the tumors originated in the adrenal glands, 13.6% (3/22) were in the paravertebral region, 9.1% (2/22) in the pelvic cavity, and 13.6% (3/22) in the chest. According to Evan's system, 50% (11/22) of the patients were stage III, 40.9% (9/22) stage IV, and 9.1% (2/22) stage IV-S. Most of the tumors were poorly defined. One half of the tumors contained calcifications. 22.7% (5/22) had invaded the spinal canal. Metastases (bone, liver, or pulmonary) were seen in CT scans in 72.7% (8/11) of the patients at stage IV or IV-S. Most of the patients only sought help when the neuroblastomas had already reached late stages. CT scans of the neuroblastomas not only demonstrated the extent of the primary tumor, but also helped to detect metastases.

INTRODUCTION

Neuroblastoma is the most common extracranial solid malignant tumor in children; it is the third most common malignancy of childhood, surpassed in incidence only by acute leukemia and primary brain tumor.[1] The patient's age, site of tumor, and stage of disease at initial diagnosis are the main factors in the prognosis of this disease. Patients under one year of age have a more favorable prognosis. Thoracic neuroblastomas have a better prognosis than abdominal or pelvic neuroblastomas.

CT and bone scintigraphy in conjunction with pertinent clinical data and bone marrow aspiration will accurately stage at least 95% of patients using Evan's classification.[2] CT is essential for the confirmation, localization, and staging of neuroblastoma.

MATERIALS AND METHODS

We reviewed CT scans performed from October 1991 to December 1997 on 22 children ranging in age from 2 months to 15 years with neuroblastoma not yet treated. The average age was about 4 years. There were 9 girls and 13 boys. The presenting symptoms were abdominal mass (10/22), bone pain (6/22), prolonged fever (2/22), neck mass (1/22), paraplegia (1/22), and periorbital ecchymosis (1/22). One child had an incidental posterior mediastinal mass.

The diagnoses were made from bone marrow aspiration biopsy, fine needle aspiration biopsy of the tumor, cervical lymph node biopsy, or histological examination after surgical removal of the tumor. Twenty-one tumors were neuroblastomas histologically. One thoracic tumor was ganglioneuroma with neuroblastoma foci.

According to Evan's system,[3] 50% (11/22) of the patients were stage III, 40.9% (9/22) stage IV, and 9.1% (2/22) stage IV-S.

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The CT scans were performed using contiguous axial 10 mm slices. Scans were performed both before and after administration of bolus intravenous contrast medium. Oral contrast medium was also given to patients with abdominal neuroblastoma. The CT scans were examined for

1. site of the main tumor mass;
2. extension of the tumor mass;
3. encasement of the great vessels (aorta and its branches, IVC, or SVC);
4. tumor characteristics (margins, calcifications, and areas of necrosis);
5. intraspinal extension; and
6. liver, bone, or pulmonary metastases.

RESULTS

It was found that of the 22 neuroblastomas, 14 (63.6%) originated in the adrenal glands (Fig. 1), 3 (13.6%) were in the paravertebral region (Fig. 2), 2 (9.1%) in the pelvic cavity (Fig. 3), and 3 (13.6%) in the chest (Fig. 4).

Most of the tumors (19/22) were poorly defined. The remaining three were well defined; two of these were stage IV-S (Fig. 5) and one was stage III (Fig. 6). Encasement of the great vessels was seen in 14 children. One half of the tumors contained calcifications. All of the tumors had areas of necrosis. 22.7% (5/22) had invaded the spinal canal; only 1 child had symptoms of cord compression. On CT scans of the primary tumor, bone metastases were seen in 4 children (Fig. 7), liver metastases in 2 children, and pulmonary metastases in 1 child.

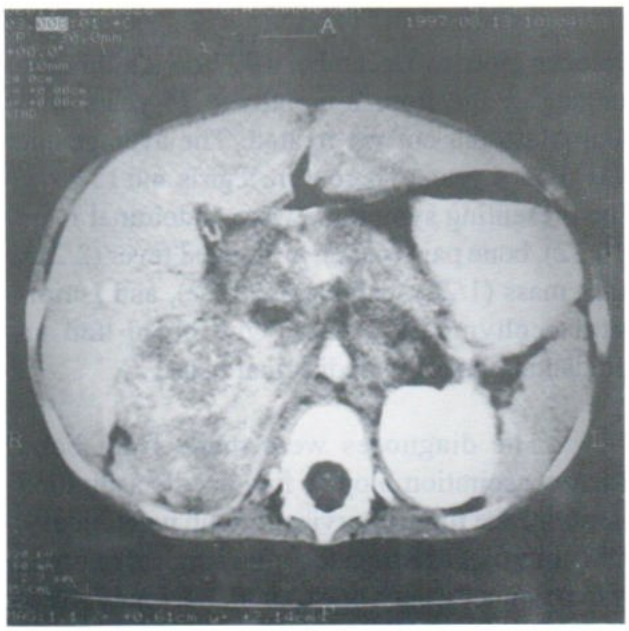

Fig. 1 Contrast CT scan shows an ill-defined inhomogeneous soft tissue mass in the right suprarenal region. The mass encases the great vessels and extends across the midline.

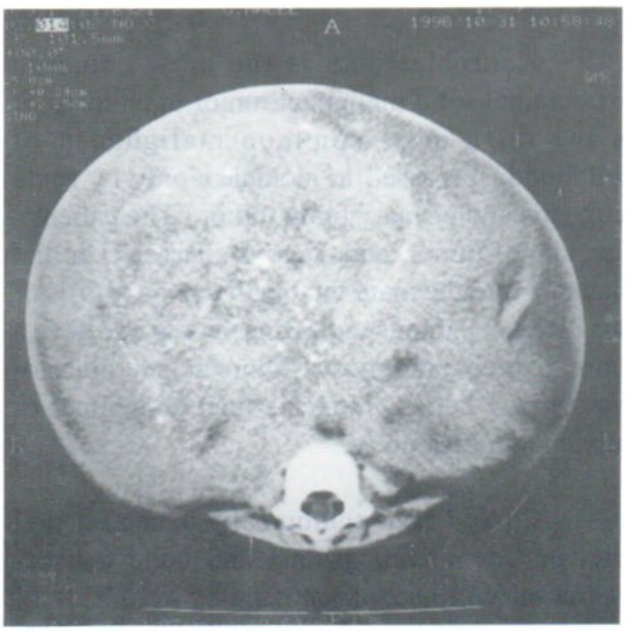

Fig. 2 Contrast CT scan shows a huge inhomogeneous soft tissue mass with multiple stippled calcifications in the paravertebral region.

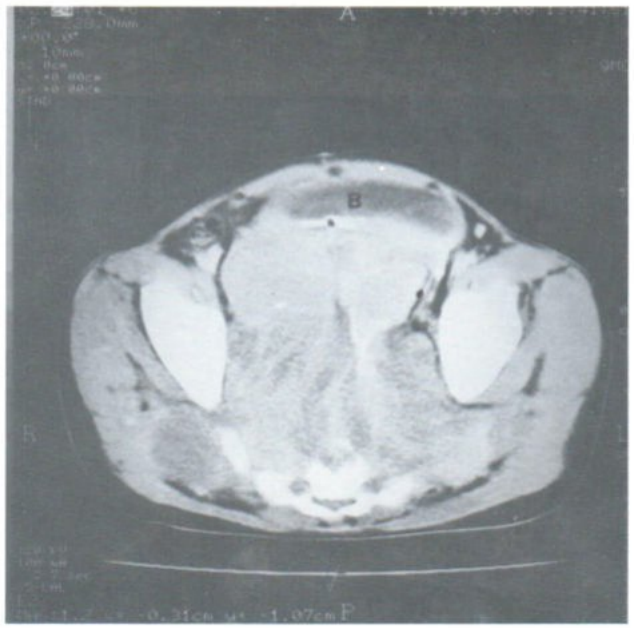

Fig. 3 Contrast CT scan shows an ill-defined inhomogeneous soft tissue mass in the pelvic cavity. The mass has extended through the pelvic side walls and destroyed the sacrum. (B= urinary bladder)

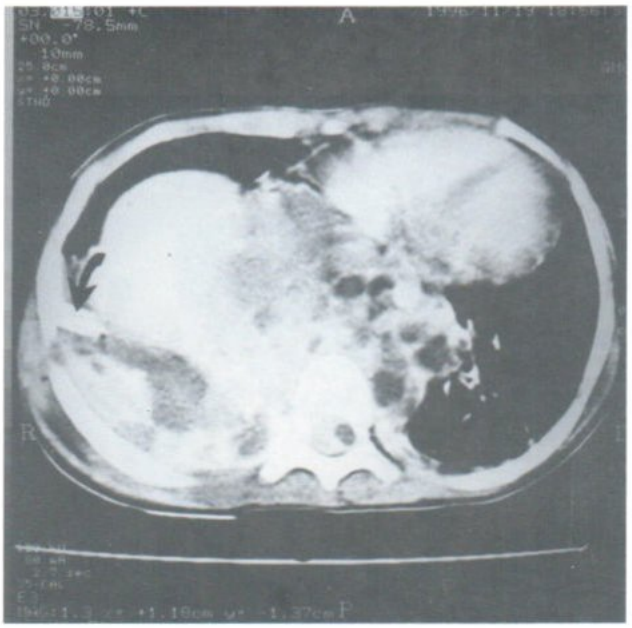

Fig. 4 Contrast CT scan shows an ill-defined inhomogeneous soft tissue mass in the mediastinum. Note the extension of the mass to the right chest wall and epidural space. (arrow = thoracostomy tube)

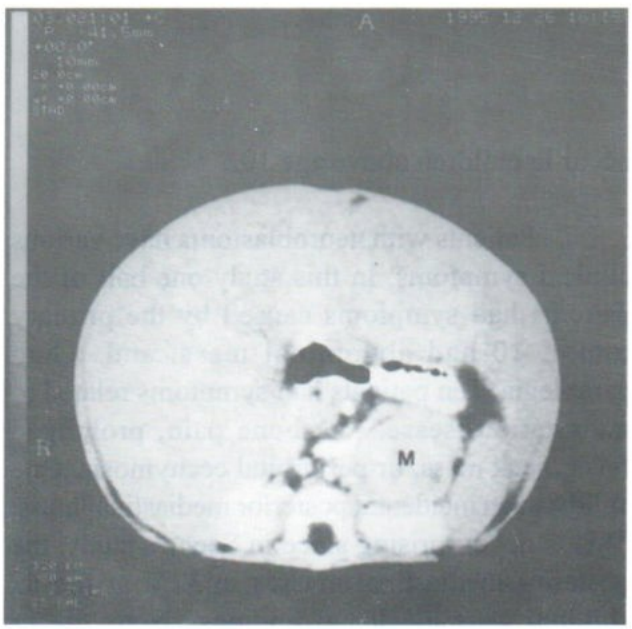

Fig. 5 Contrast CT scan shows a well-defined left suprarenal mass (M) and diffuse inhomogeneous density of the liver caused by metastases.

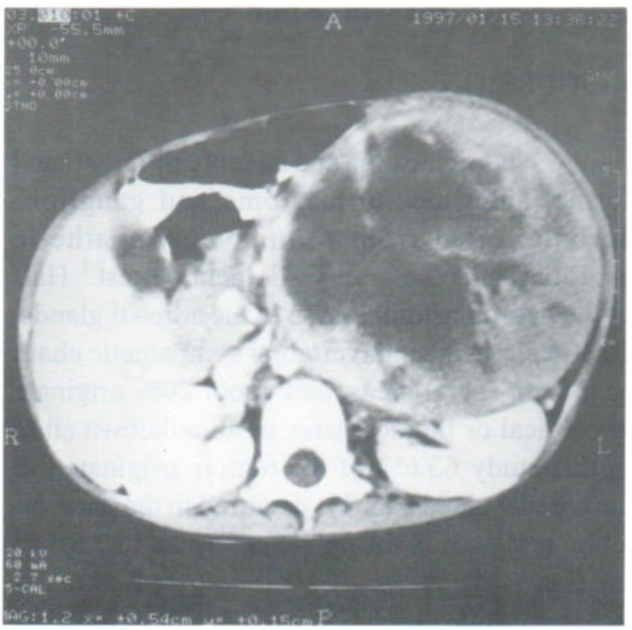

Fig. 6 Contrast CT scan shows a well-defined inhomogeneous soft tissue mass which is pressing the anterior aspect of the left kidney. It extends across the midline but does not encase the great vessels.

Fig. 7 Neuroblastoma with bone metastases

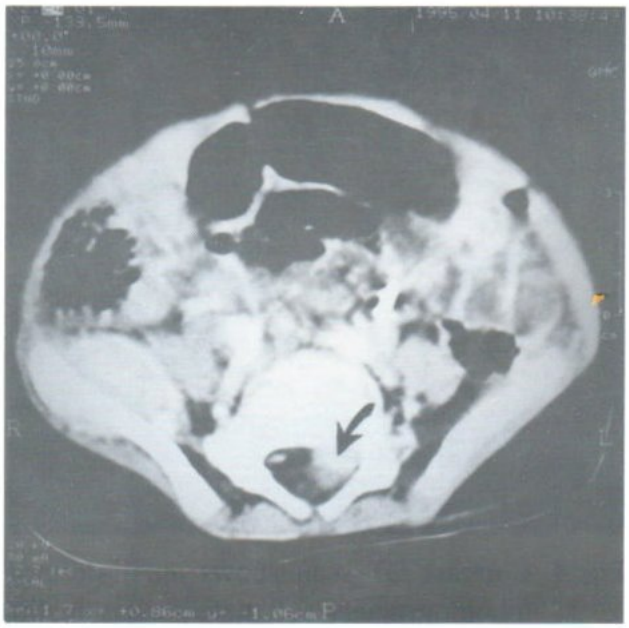

Fig. 7A Contrast CT scan shows soft tissue masses on both sides of the right iliac bone. Note the extradural mass (arrow).

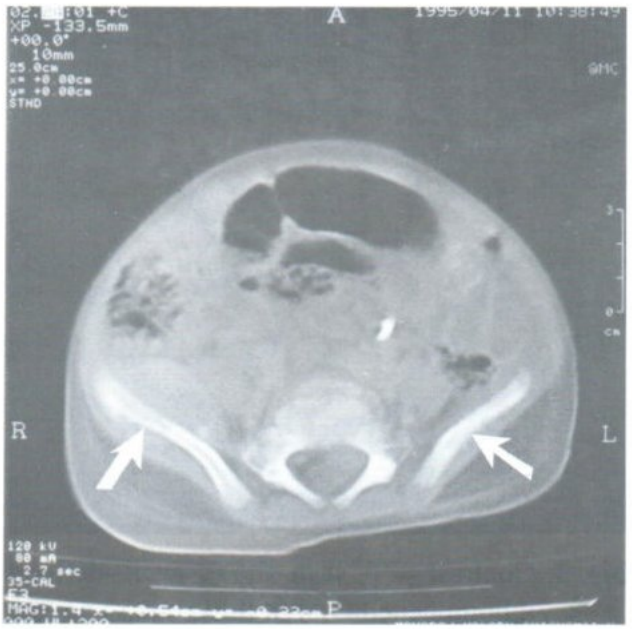

Fig. 7B CT scan (bone window display) shows destruction of the right and left iliac bones.

DISCUSSION

Neuroblastoma and its more differentiated forms, ganglioneuroblastoma and ganglioneuroma, arise from primitive sympathetic neuroblasts of the embryonic neural crest.[1] Half of the neuroblastomas arise in the adrenal glands, 30% occur in the paravertebral sympathetic chain or the pre-sacral areas, and about 20% originate in cervical or thoracic sites or an unknown site.[4] In this study 63.6% of the tumors originated in the adrenal glands, 22.7% occurred in the paravertebral region or in the pelvic cavity, and 13.6% in the chest.

The tumor is more common in boys than in girls; the male/female ratio is 1.2:1 in most large studies.[4] 75% of patients are less than 4 years of age, and fewer than 10% of neuroblastomas occur in children above age 10.[1]

Patients with neuroblastoma have various clinical symptoms. In this study one half of the patients had symptoms caused by the primary tumor, 10 had abdominal mass, and 1 had paraplegia. Ten patients had symptoms related to metastatic disease, i.e., bone pain, prolonged fever, neck mass, or periorbital ecchymosis. One child had an incidental posterior mediastinal mass. This is not surprising since in Saenz's study, the posterior mediastinal masses in 32% of the 63 children were incidental findings.[5] None of our patients had paraneoplastic syndromes, i.e. myoclonic encephalopathy of infancy;[6] syndrome of intractable watery diarrhea, hypokalemia, and achlorhydria;[1] and excessive catecholamine

syndrome (hypertension, headache, flushing).[4]

Neuroblastoma is characterized by a calcified suprarenal mass or pararenal mass.[2,7] In Peretz and Lam's experience the most reliable sign of abdominal neuroblastoma is the displacement of the IVC and aorta, particularly when they are separated from the vertebral bodies.[8] In their study calcifications were found in CT scans in 76% of patients with abdominal neuroblastoma. Golding, et al. found that 24/33 (72.7%) of thoracic and abdominal neuroblastomas contained minor or heavy calcifications.[9] We found fewer calcifications than previously reported. Only one half of our patients had calcifications in CT scans.

As most abdominal neuroblastomas arise in the adrenal gland near the aorta, celiac axis, and/or superior mesenteric artery, it is not surprising that tumors crossing the midline tend to involve vital vessels. Since surgery currently plays a major role in the treatment, the relation of the tumor to the great vessels is a more reliable and important factor in predicting the outcome of these children than the extension and location of the tumor with reference to the midline.[10] CT after intravenous contrast enhancement is helpful in assessing the great vessels. One of our stage III patients had a well defined tumor crossing the midline but not encasing the aorta or great vessels. It was able to be totally resected.

A small number of children with neuroblastoma present with symptoms of cord compression.[11] Golding et al found that a significant number of children had clinically unsuspected extensions of tumor into the spinal canal[9]; four of our patients with such extensions had no clinical signs.

Common sites of metastases for neuroblastomas are skeleton, bone marrow, liver, lymph nodes, and skin. The pattern of metastases varies greatly with age. Bone metastases are extremely common in children over one year of age, usually involving the long bones and orbits. In newborn liver and skin metastases are more common than bone metastases.[1] Pulmonary parenchymal metastases are considered extremely rare in patients with neuroblastomas. Pulmonary metastases occur in patients with widespread disseminated disease. It is a grave prognostic sign and considered a pre-terminal event.[1]

Ultrasonography is usually the initial imaging modality for a child with a palpable abdominal mass. It has limited value in detecting retroperitoneal and retrocrural lymph node metastases. It is unable to detect extradural extension of tumor into the vertebral canal.

CT is extremely valuable for determining the extent of neuroblastomas. It is able to detect prevertebral extension of tumor across the midline, encasement of the great vessels, renal invasion, nodal metastases, intraspinal extension, liver metastases, response to therapy, and tumor recurrence. It is excellent for demonstrating retrocrural extension to the chest, which commonly occurs in abdominal neuroblastoma.

MR imaging plays an important role in defining tumor resectability; it is better than CT in defining vascular encasement, hepatic metastases, bone marrow metastases, and intraspinal extension.[12-14] The disadvantage of MRI is that bone erosion and pulmonary metastases are less well visualized than they are with CT.[5]

CONCLUSIONS

Most of our patients only sought help when the neuroblastomas had already reached late stages. CT scans of most of our stage IV and IV-S tumors not only demonstrated the extent of the primary tumor, but they also helped to detect metastases (bone, liver, or pulmonary).

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